Ectopia cordis thoracalis with cystic hygroma, syndactyly and cleft lip and cleft palate

R. K. Ghritlaharey, Gaurav Gupta, A. S. Kushwaha, R. Chanchlani
Department of Pediatric Surgery, Gandhi Medical College and Associated, Kamla Nehru and Hamidia Hospitals Bhopal - 462 001, MP, India

Correspondence: Dr. Rajendra K. Ghritlaharey, Department of Pediatric Surgery, Gandhi Medical College and Associated Kamla Nehru and Hamidia Hospitals, Bhopal - 462 001, MP, India. E-mail: drrajendrak1@rediffmail.com

CASE REPORT

A full term female baby of 2.3 kg was born to a Gravida I Para 0, 22-years-old mother on June 12, 2006. She was brought to us one hour after birth. The pregnancy and hospital delivery were uneventful. There was no history of infection, intake of any teratogens, drugs or exposure to radiation, etc. in antenatal period.

Clinical examination revealed peripheral cyanosis with heart rate of 146/min and respiratory rate of 48/min. Her heart was lying outside the thoracic cavity and apex of the heart was pointing anterio-superiorly. The heart had pericardial covering and coronary and few great vessels could be seen [Figure 1]. She also had associated right-sided complete cleft lip and cleft palate, cystic hygroma at right side neck of 4 x 3 cm in size. Syndactyly of little, ring, and middle fingers of right hand also present [Figure 2]. Investigations revealed no diaphragmatic hernia. After mobilization of skin around the heart, it was easily repositioned within the thoracic cavity and skin closure was done. She died 2 hours after operation due to cardiac arrest. We presumed that our patient also had intrinsic cardiac defects, although neither echocardiography nor post-mortem examination was done.

DISCUSSION

Ectopia cordis (EC) is defined as complete or partial displacement of the heart outside the thoracic cavity. It is a rare congenital defect in fusion of the anterior chest
wall resulting in extra thoracic location of the heart. The estimated prevalence of ectopia cordis is 5.5-7.9 per million live births and may occur more in females.\(^{1,2}\)

Depending upon the location of the heart, it could be classified into five types: cervical, cervicothoracic, thoracic, thoracoabdominal, and abdominal.\(^{3,5}\) Thoracic and thoracoabdominal EC accounts for about 85% of the cases. Thoracic EC is a classic naked heart with no overlying somatic structures.

EC can be diagnosed by routine prenatal ultrasonography as early as in 10-12 weeks of pregnancy.\(^{3,4}\) Most cases of EC result in stillbirth or die shortly after birth. It is frequently associated with other intrinsic cardiac as well as other congenital defects involving multiple organ systems.\(^{4,5}\)

Surgical correction of these defects is complex and generally requires a staged closure as:
1. Covering of the naked heart by skin
2. Placement of the heart into the thoracic cavity
   Correction of the intrinsic cardiac defects is desirable prior to orthotopic location of heart
3. Sternal/thoracic reconstruction

Haller first described the term ectopia cordis in 1706. The first attempted repair of EC was performed in 1925 by Cutler and Wilens; however, it was Koop who achieved first successful repair of thoracic EC in two stages in 1975. Amato et al. reported successful single stage repair of thoracic EC in 1995.\(^{6,7}\)

Review of literature reported only three survivors of 29 attempted repairs of thoracic EC, to which few more unsuccessful attempts have been added.\(^{8-10}\) Successful repair of such anomalies is dictated by the presence and severity of the intrinsic cardiac defects and associated other congenital anomalies, rather than the type of surgical approach itself.

Advances in the fetal ultrasound techniques have aided in the early recognition of such anomalies and termination of pregnancy prior to viability should be considered and discussed with parents.

REFERENCES


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